



## Overview

### Points To Remember About Autoinflammatory Diseases

- Autoinflammatory diseases refer to problems with the immune system, which usually fights off viruses, bacteria, and infection. It results when your immune cells attack your body by mistake.
- Autoinflammatory diseases can cause fever, rash, swelling of joints and other tissues, and serious buildup of a blood protein in organs.
- In most cases, your doctor will prescribe medications to reduce pain and swelling or that act on the immune system.

Your immune system is the network of cells and tissues throughout your body that work together to defend you from viruses, bacteria, and infection. It tries to identify, kill, and eliminate the invaders that might hurt you.

Parts of the immune system include:

- **Acquired (or adaptive) immune system**, which develops as you grow. Invaders provoke your body into producing antibodies so that your body “remembers” those invaders. Your body can then fight them if they come back.
- **Innate (or inborn) immune system**, which uses white blood cells instead of antibodies to destroy invaders.

### What happens in autoinflammatory diseases?

Autoinflammatory diseases refer to problems with the innate immune system's reactions. Immune cells target the body's own healthy tissues by mistake, signaling the body to attack them. This can cause intense episodes of inflammation that result in such symptoms as fever, rash, or joint swelling. These diseases also carry the risk of amyloidosis, a potentially fatal

buildup of a blood protein in vital organs.

## **Types**

Some examples of autoinflammatory disease and their symptoms include:

- **Familial Mediterranean Fever (FMF)**, which causes recurring bouts of fever. Other symptoms include:
  - Severe abdominal pain due to inflammation of the stomach cavity (peritonitis).
  - Arthritis (painful, swollen joints).
  - Chest pain from inflammation of the lung cavity (pleurisy).
  - Skin rashes.
- **Neonatal Onset Multisystem Inflammatory Disease (NOMID)** affects the skin, joints, eyes, and central nervous system. For most children, the first sign of the disease is a rash that develops within the first six weeks of life. Other problems can follow, including:
  - Fever.
  - Meningitis.
  - Joint damage.
  - Vision loss.
  - Hearing loss.
  - Mental retardation.
- **Tumor Necrosis Factor Receptor-Associated Periodic Syndrome (TRAPS)** is associated with:
  - Long, dramatic, episodes of high fever.
  - Severe pain in the abdomen, chest, or joints.
  - Skin rash.
  - Inflammation in or around the eyes.
- **Deficiency of the Interleukin-1 Receptor Antagonist (DIRA)** can cause the following serious conditions in children:
  - Swelling of bone tissue.
  - Bone pain and deformity.
  - Inflammation of the layer of connective tissue around bone.
  - Skin rash that can cover most of the body.
- **Behçet's Disease** can cause the following symptoms:
  - Mouth or genital sores.
  - Redness and swelling in the eyes.
  - Arthritis.
  - Skin problems.
  - Swelling of the digestive system, brain, and spinal cord.
- **Chronic Atypical Neutrophilic Dermatitis With Lipodystrophy and Elevated Temperature (CANDLE)**
  - Recurrent fevers.
  - Purpura.
  - Joint pain, contractures, and developmental delay.
  - Facial changes, including a loss of fat on the face, and swollen lips and eyelids.

## Causes

Autoinflammatory diseases are typically caused by changes in certain genes. This causes problems with proteins that are important in specific body functions. For some diseases, the cause is unknown.

## Treatment

Treatments for autoinflammatory diseases can involve medications that:

- Reduce inflammation.
- Suppress the immune system.

## Research Progress

Researchers continue to explore the genetics and causes of autoinflammatory diseases. Understanding the causes of these diseases will improve both diagnosis and treatment.

## Related Resources

### **U.S. Food and Drug Administration**

Toll free: 888-INFO-FDA (888-463-6332)

Website: <https://www.fda.gov>

Drugs@FDA at <https://www.accessdata.fda.gov/scripts/cder/daf>. Drugs@FDA is a searchable catalog of FDA-approved drug products.

### **Centers for Disease Control and Prevention, National Center for Health Statistics**

Website: <https://www.cdc.gov/nchs>

### **National Institute of Allergy and Infectious Diseases**

Website: <https://www.niaid.nih.gov/diseases-conditions/autoinflammatory-diseases>

### **American Autoinflammatory Related Diseases Association (AARDA)**

Website: <https://www.aarda.org>

If you need more information about available resources in your language or other languages, please visit our webpages below or contact the NIAMS Information Clearinghouse at [NIAMSInfo@mail.nih.gov](mailto:NIAMSInfo@mail.nih.gov).

- [Asian Language Health Information](#)
- [Spanish Language Health Information](#)

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## **Related Information**

[¿Qué es el síndrome de Behçet? \(Esenciales: hojas informativas de fácil lectura\)](#)

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[Autoinflammatory Diseases, Understanding Behçet's Disease, Easy-to-Read Fast Fact](#)  
[Behçet's Disease, Questions and Answers about](#)